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A unique inside look in to the life of a CF patient: from the patient and RRT perspective

Abstract

As a Cystic Fibrosis patient and an RRT, I have a unique perspective. Not only do I treat patients for a living, I am a patient. In this unique article, I'll talk about my life as a CF patient. I'll discuss the struggles that CF patients, their spouses, and family members face on a daily basis. My goal as a CF patient is to give you the reader a back stage pass if you will, into my life. I hope to provide you with a better understanding of what CF looks like from a patient standpoint. I want the healthcare professional to understand the diversity in the way CF patients are affected by the disease. I'll expose and dispel myths about CF, talk about ways CFers are labeled the importance of coping with a chronic illness, and what it takes to combat the disease. My goal as a respiratory therapist and fellow healthcare worker is to inspire you to give the best care you can to your patients. I'll talk about treatment options, cost of managing CF, share statistics, and all sorts of things related to CF. Most importantly, I will share my testimony of the impact healthcare workers have on their patients.

Keywords: cystic fibrosis, CF, CFer (CF patient)

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Opinion

My name is Jeremy. I'm a respiratory therapist and I have Cystic Fibrosis. Being a CF patient and RRT gives me a very unique perspective of not only the respiratory profession but healthcare in general. I have given several presentations titled "An inside Look into the Life of a CFer." The presentation addresses the various struggles CF patients' face, how important the role of RT is in our care and what it is like to be a patient. My hope is that this article will do the presentation justice.

I was unofficially diagnosed with CF at birth and officially diagnosed at 2years of age. I had an older sister, Christa, who my parents lost around 8 months old to CF and sepsis. Christa was the first in my family tree to be diagnosed with CF and it was because of her, doctors knew to test my younger sister, Kara and I. Kara does not have CF. She does however only have one lung, but that's another story entirely.

Cystic Fibrosis is a hereditary disorder affecting the exocrine glands. It causes the production of abnormally thick mucus, leading to the blockage of the pancreatic ducts, intestines, reproductive tracts and bronchi often resulting in respiratory infection. Now, if you were to tell someone with CF (a CFer) they have a disease called Cystic Fibrosis and rattled off the above definition. You would likely get a resounding "WHAAAAAAT?!?!" The textbook definition of CF means nothing to us CFers. So what does Cystic Fibrosis mean to us?

What CF means to us is determined by how it affects our everyday lives. For example, from birth until around 13years of age, CF meant virtually nothing to me. It was a label I carried. CF meant I had to take treatments (that I never took), go to the doctor and have breathing tests, chest x-rays, blood draws and was asked to cough something into a cup, which I rarely could do. I could outrun every kid I knew. I played soccer, baseball and basketball. I was virtually unaffected. Between the ages of 14-17, I noticed I kept getting "colds" twice a year when the seasons would change and had to take a "Z-pack." Each year they seemed to last longer and longer. I wondered if they might be related to my "CF" but brushed it off as a coincidence. At 17 I made the mistake to beat all mistakes. I started smoking. I enjoyed the buzz from it and liked watching the smoke as I blew it out of my lungs. It was the cool thing to do. I was very under-educated about CF and its effects on the lungs. I blame myself. My parents and physicians tried to educate me and get me to take treatments. I didn't listen because I didn't notice anything was wrong. I was healthy, and stayed healthy despite not doing any of the treatments. In my mind, I didn't need them. Subsequently at age 18, I had my first hospital admission. Go figure right? Another admission followed a year later. Having to be hospitalized scared me to death. From that age on I began taking better and better care of myself as I watched the disease affect me more and more. With each passing year, what CF means to me changes as it progresses. I've had to start doing more treatments and struggle to keep my lung function up. CF is constantly progressing and so does our view of it. As far as my knowledge of CF, I did a research project in college for a composition class about Cystic Fibrosis. I thought it would earn me sympathy "A." It worked! At the same time I learned a great deal about CF.

Cystic Fibrosis is a lifestyle disease. By that, I mean your lifestyle has to be molded around the disease. This is crucial. If you are unable to do this, you will not cope with the disease. If you can't cope with it, it will overtake you, and you will lose your battle far sooner than you should have. I have an amazing support system. They help me cope tremendously. I have family and friends that care and help to encourage and motivate me. This is not the case for all CFers. Some have absolutely zero support outside the hospital walls. For them it is extremely tough to find reasons to stay motivated to fighting this terrible disease. Beyond my support system, there are several ways I mold my lifestyle to fit around CF. I work 8 hour shifts instead of 12's. Most people think that's crazy. When I worked 12hour nights I never took treatments when I worked more than one shift in a row. I was too tired. Eight hour shifts work for me. They give me plenty of time to do my treatments when I get home and still have a life. I try to get 8hours

of sleep a night. My doctors stress that I need 8hours because my body is constantly fighting infection and burning energy. I take pills every day with every meal. CFers are deficient in the vitamins A,D,E and K which are fat soluble vitamins. Due to our pancreatic insufficiency, which we take enzymes for every time we eat, we cannot absorb those vitamins and must supplement them. I take two sets of treatments a day for a total of around 2hours minimum, every day. I get up at 4:45am before work and do a set, then do another set when I come home. With CF, exercise is a necessity. If I do not exercise and do two sets of treatments a day, my PFT's will drop. I have seen it happen when I slack on my routine. My doctors can tell also. They know when I come in with a dip in lung function but look good and are not having an exacerbation; I must be slacking in my exercise. It may sound like a lot of work, and truthfully it is. Keeping up with the beast inside known as "Cystic Fibrosis" is tough and takes a lot of determination and hard work. However, CF does not have to limit the quality of your life. We all know at the end of the disease process it will, but for most of us, we have a lot of good years to live, virtually uninhibited, before that happens.

CF is mentally, physically and emotionally draining. I get tired of having to take treatments. Especially when there's something going on with my family or friends and I have to show up late or miss out because of treatments. I get tired of coughing. When I get sick or have an exacerbation, I cough like crazy. Lying down to sleep is an ordeal because I can't stop coughing when I lay flat. My CF physician told me of a study he read about the number of times a person with CF coughs each day. Can you guess? The average CFer, when they are not sick, coughs around 600times a day. That's a lot of coughing. It can get old. We get tired of facing the mental and emotional struggles. Movies like P.S. I Love You where the lead character dies young and leaves his young wife a widow is reality checks for us. CF is a terminal illness that fights to destroy our bodies 24/7, 365 days a year. It's relentless. There are no timeouts or breaks. We have to fight often and fight hard. It can be wearying at times. The disease could take a turn and begin progressing extremely fast, and we could be gone within a year. We just plain get tired. The average CFer (with a chronic pseudomonas infection) walks around with 2.5million CFU's (colony forming units) of pseudomonas in their lungs when they are "healthy" and around 10million CFU's when they are having an exacerbation. Our body is constantly burning energy, trying to eliminate the infection, which is a never-ending battle. It leaves us exhausted from our body burning through so much energy. This is one of the reasons so many CF patients can eat insane amounts of food and yet gain no weight. Lastly we know someday no matter how hard we fight, we will lose the battle. CF is terminal.

Parents of CFers also face many struggles. They are the sole care-givers for a child with a terminal illness until the child becomes old enough to take on that responsibility themselves. They are tasked with making sure treatments get done, medicines get ordered, and doctor appointments are made and kept, educating their child about the disease and have the often large financial burden CF brings as well. Raising a child is stressful and a full time job in and of itself. Throw in a terminal illness and it can become over-whelming to say the least. CF parents are very special and deserve a lot of credit for the burdens they carry. CF parents often carry a great deal of guilt for "giving" their child CF. I was unaware of this until my dad told me my mom had been carrying this guilt. She felt as though it was her fault I had to go through the hardships CF brings and in order to have children of my own would have to seek fertility help. She felt as though she "gave" me CF. Many CF parents have a tendency to spoil their children as a result of this guilt. What parent doesn't want to spoil their kids though? However, as parents of CFers there comes a point where they have to let their child take responsibility for their disease. It's part of coping. If they don't they may be stuck in my next point the rest of their lives.

There is a phase I think every CFer goes through. I call it the "Pity-Party" phase. We feel sorry for ourselves. We wonder "Why me?" We look at people around us with jealousy and frustration that they don't have to take treatments and spend so much time struggling to stay healthy. Some of us will live our entire lives in this phase. It's unhealthy. It is important to accept the disease for what it is and understand that we cannot change it. We have to make the decision to fight the disease or sit back and watch as it kill us. My decision was the latter.

I was not bitter. I had accepted CF for what it was terminal. I was ready to go to heaven and be with my sister Christa. To meet her for the first time. To be rid of the disease Cystic Fibrosis and spend eternity with God. I didn't want to give up my time fighting a battle I knew I would lose. It seemed foolish when I already knew the outcome. I could spend that time having fun and enjoying life instead of being strapped to a machine and breathing medicine all the time. A lot of CFers make this decision for the same reason I did, and sadly, a lot of them never change their mind. Fortunately, I did.

I met my wife. She changed my mind. My wife gave me a new reason to fight the disease. My goal now is to live as long as I can for her. To give her the best life I possibly can for choosing to love me in spite of my CF. I have been blessed with an amazing wife! In April of 2013 I was given two more reasons to fight. My wife and I had twin boys via in vitro fertilization. Whoo-hoo!

CF spouses face many struggles of their own. If you're afraid of commitment marry a CFer. Let me clarify that statement. I say that not because we will die young. It takes an extremely special and dedicated person to marry someone with CF. There are a lot of hardships. The first being financial. The average cost of managing CF in 2011 was $48,000. If insurance covers 80%, you're left with $9,800 out of pocket. Most of us opt to have a lung transplant if we're lucky enough to receive one. The average cost of a bilateral lung transplant in 2011 was $800,000. Again, after insurance's 80%, you're left between $150,000-$180,000 out of pocket. 98% of men with Cystic Fibrosis are infertile because of a lack of or blockage of the vas deferens. That means the only option for having children of our own is via in vitro fertilization (IVF). This costs around $19,000 and is no guarantee. It's about a 50% chance it will work. Lastly, they have no idea how long they will have with us. The median life expectancy made it into the 40's this year. However, 10% of newborn cases still die before the age of one, and I can't count how many I've known that have passed in their early 20's. There's always a chance of death for everyone. If anyone knows this better, it's healthcare workers. But, it's different when you are waiting, knowing someday your loved one is going to lose the battle.

I mentioned earlier that CF was a label I carried. As CFers we carry labels with us everywhere we go. We are contagious, we cannot have children, we will all die before we turn 20, and we're sickly and weak are just a few I've personally witnessed. People hear our cough and act like we have the bubonic plague. I asked my CF doctor about this. I asked him if I could make my wife and children sick. He said

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no. He said they will make me sick. They may catch a simple cold that I'll catch and as a result, may need to be admitted for IV antibiotics. The same goes with the general public. When I was researching CF for the paper I did in college. I read that 98% of men with CF can't have kids. It was devastating. I had always dreamed of having several children. It was a true statement but misleading. Modern science has ways of making it possible. IVF, as I mentioned earlier, makes it possible for CF men to have families of their own. This was a big deal when my wife and I began to talk marriage while we were still dating. My mother-in-law had read that same fact and was worried for her daughter. She knew how much my wife wanted a family of her own. It's not impossible. It just takes a lot more effort and money than for most people. The worst I have ever been labeled was during a surgery I had for a bowel obstruction when I was 21. The surgeon repairing the bowel obstruction deliberately did not reattach nerves to my stomach muscles or take the time to stitch me up strongly and nicely. I was left with stomach muscles that I still cannot tighten and a gnarly looking scar (I'm a guy so it's ok. Girls dig scars right?). He labeled me as having "a poor prognosis of life" in his operative note and therefore intentionally did not take the time to finish the job right. He was uneducated to the truth of my health at the time and to the advances in CF care. A lot of people are which is why I am so adamant about CF awareness. I don't want to see younger CFers be labeled as I have because people are uneducated. Which leads me to the last label? That we are all sickly and weak. When we hear the word terminal disease, it's our human nature to picture someone frail and on the verge of death. As CFers, most of us live very normal lives and are fully able to participate in anything we want. Being active is part of keeping ourselves healthy. For example, I've been skydiving with no problems. I go snowboarding in the mountains in Colorado every year and have no issues with the altitude. I wake board every summer with my wife's family at the lake. I've been SNUBA diving (a form of SCUBA diving) twice with no issues (although I do have a funny story about that from my wife and I's honeymoon). I went on a 30minute hike up a mountain in the jungles of St. Lucia to zip-line back down. We live very normal lives. Our lung function may be significantly lower than those around us, but we can keep up as if it isn't. Most the time people see us and would never guess we have a terminal illness.

Lastly I will leave you with this. Take the time to care. It's in the name of our field, healthcare. No one wants to spend 45minutes with an RT who has a bad attitude, be stuck with a nurse all day who's in a foul mood, or any other healthcare worker. As members of the healthcare team, you are invaluable in the care of a CFer.

"Being healthy simply means you're dying at the slowest rate possible."

As CFers this is our goal. To slow the progression of the disease as much as possible. When we are admitted, we're there to gain back lung function that we desperately don't want to lose forever. Lung function =years for us. You're adding years to our lives by helping us get better. Make the most of the time with your patients. You could make a difference in their lives. You could be the reason a stubborn CFer starts taking care of them. All because you took the time to get to know them beyond their disease and care. I have my own real life hero. A guy named G.W. He was one of my RT's at my first hospital admission. He later became my supervisor, at my first job after graduating RT school. He has since become a lifelong friend. From the first time we met he took the time to care. He invested his time into getting to know me and has been there through the years to encourage me, motivate me, and give me the kick in the butt I sometimes need. You could be someone's G.W.!

To all the healthcare employees reading this I would like to thank you on behalf of the CF community. Thank you for tolerating our unusual behaviors, our CF quirks, for beating on us (for those who still do manual CPT), for taking the time to get to know us beyond our disease and thank you for caring! We really do appreciate it (even though we may not always show it).

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